Implementing phenotype-based risk stratification of BAV aortopathy in surgical practice

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Disclosure of Interest

Speaker name: Evaldas Girdauskas

- I do not have any potential conflict of interest
Bicuspid aortopathy

- Prevalence 50 – 70 %
- Heterogeneity in BAV aortopathy (*BAV phenotypes*)
  - BAV + dilatation of tubular ascending aorta
  - BAV + root dilatation (Root phenotype)
- Normal-sized aorta in BAV
Phenotypic classification of aortopathy
BAV phenotypes & outcome
BAV insufficiency was associated with a 10-fold higher risk of post-AVR aortic dissection compared with BAV stenosis (OR: 10.0; 95% C.I. 6.2-16.2; p < 0.001)

Histological aortic wall lesions

BAV root phenotype
Aortic events after isolated aortic valve replacement for bicuspid aortic valve root phenotype: echocardiographic follow-up study

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CONCLUSIONS: Patients with a BAV root phenotype are at significant risk of aortic events after isolated AVR.
Aortopathy gene panel
- 20 candidate genes
- NGS analysis

likely / potentially pathogenic variants in 19 / 63 (30%) patients with BAV root phenotype

will be presented at 30th EACTS Annual Meeting, October 2016, Barcelona
BAV root phenotype
Risk of late aortic events after an isolated aortic valve replacement for bicuspid aortic valve stenosis with concomitant ascending aortic dilation

Evaldas Girdauskas, Kushtrim Disha, Heinrich H. Raisin, Maria-Anna Secknus, Michael A. Borger and Thomas Kuntze

OBJECTIVES: The optimal surgical treatment of patients with bicuspid aortic valve (BAV) disease and ascending aortic aneurysm is controversial. The aim of this study was to evaluate the risk of late aortic events after an isolated aortic valve replacement (AVR) for BAV stenosis with concomitant mild-to-moderate proximal aortic dilation.

METHODS: A retrospective analysis was performed on 153 consecutive patients (mean age 54.2 ± 10.5 years, 73% men) with BAV and ascending aortic aneurysm (>40 mm) who underwent AVR from 1995 to 2000. The mean follow-up (1759 patient-years) was 11 years. Simultaneous ascending aortic surgery was performed in 97 and 94%, respectively, at 10 and 15 years. Actuarial freedom from adverse aortic events was 95% at 10 years and 93% at 15 years.

RESULTS: Actuarial freedom from adverse aortic events was 95% at 10 years and 93% at 15 years.

CONCLUSIONS: BAV patients with aortic valve stenosis and concomitant mild-to-moderate ascending aortic dilation are at a considerably low risk of adverse aortic events at 15 years after an isolated AVR. The BAV phenotype should be considered when determining the risk of subsequent adverse aortic events and the need for concomitant aortic replacement.

- Focus on clinically overt events (SCD, redo's, dissection)
- No systematic post-AVR imaging of proximal aorta

Mid-ascending phenotype

The fate of mild-to-moderate proximal aortic dilatation after isolated aortic valve replacement for bicuspid aortic valve stenosis: a magnetic resonance imaging follow-up study†

Evaldas Girdauskas*, Mina Rouman*, Kushtrim Disha*, Georg Dubslaff*, Beatrix Fey*, Martin Misfeld†, Kambis Mashayekhi*, Michael A. Borger* and Thomas Kuntze*

Mid-ascending phenotype
BAV & normal-sized aorta
BAV & normal-sized aorta
Are normal-sized ascending aortas at risk of late aortic events after aortic valve replacement for bicuspid aortic valve disease?

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Table 3: Causes of late deaths

<table>
<thead>
<tr>
<th>Cause of death</th>
<th>BAV-AS (n = 173)</th>
<th>BAV-AR (n = 27)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cardiac death</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Congestive heart failure</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Pulmonary embolism</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Valve-related haemorrhage</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Sudden death</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>Non-cardiac death</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Malignancy</td>
<td>7</td>
<td>1</td>
</tr>
<tr>
<td>Chronic end-stage disease</td>
<td>6</td>
<td>1</td>
</tr>
<tr>
<td>Infection</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Peripheral arterial vascular disease</td>
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<td></td>
</tr>
<tr>
<td>Acute multiorgan failure</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Total deaths</td>
<td>22</td>
<td>3</td>
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Proximal aortic phenotype

<table>
<thead>
<tr>
<th>Aortic phenotype</th>
<th>BAV-AS</th>
<th>BAV-AI</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal aorta</td>
<td>61 (44)</td>
<td>10 (29)</td>
<td>0.1</td>
</tr>
<tr>
<td>Root dilatation</td>
<td>10 (7)</td>
<td>10 (29)</td>
<td>0.01</td>
</tr>
<tr>
<td>Mid-ascending</td>
<td>63 (47)</td>
<td>15 (42)</td>
<td>0.7</td>
</tr>
<tr>
<td>Distal ascending / arch</td>
<td>3 (2)</td>
<td>0 (0)</td>
<td>0.8</td>
</tr>
</tbody>
</table>
Thank you!